

Connective Tissue and Musculoskeletal  
Diseases (ERN ReCONNECT)

## REVIEW

Idiopathic inflammatory myopathies: state  
of the art on clinical practice guidelines

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**ABSTRACT**

Idiopathic inflammatory myopathies (IIMs) encompass a heterogeneous group of rare autoimmune diseases characterised by muscle weakness and inflammation, but in antisynthetase syndrome arthritis and interstitial lung disease are more frequent and often inaugurate the disease. Clinical practice guidelines (CPGs) have been proposed for IIMs, but they are sparse and heterogeneous. This work aimed at identifying: i) current available CPGs for IIMs, ii) patients' and clinicians' unmet needs not covered by CPGs. It has been performed in the framework of the European Reference Network on rare and complex connective tissue and musculoskeletal diseases (ReCONNECT), a network of centre of expertise and patients funded by the European Union's Health Programme. Fourteen original CPGs were identified, notably recommending that: i) extra-muscular involvements should be assessed; ii) corticosteroids and methotrexate or azathioprine are first-line therapies of IIMs. iii) IVIG is a treatment of resistant-DM that may be also used in other resistant-IIMs; iv) physical therapy and sun protection (in DM patients) are part of the treatment; v) tumour screening for patients with DM include imaging of chest, abdomen, pelvis and breast (in woman) along with colonoscopy (in patients over 50 years); vi) disease activity and damages should be monitored using standardised and validated tools. Yet, only half of these CPGs were evidence-based. Crucial unmet needs were identified both by patients and clinicians. In particular, there was a lack of large multidisciplinary working group and of patients' preferences. The following fields were not or inappropriately targeted: diagnosis; management of extra-muscular involvements other than skin; co-morbidities and severe manifestations.

**Key messages****What is already known about this subject?**

► Clinical practice guidelines (CPGs) have been proposed for idiopathic inflammatory myopathies (IIMs) but no review of "what we have" and patients' as well as clinicians' unmet needs has been performed so far.

**What does this study add?**

► Fourteen original CPGs were identified, covering important issues. Yet, only half of these CPGs were evidence-based, and crucial unmet needs were identified both by patients and clinicians.

**How might this impact on clinical practice?**

► Future CPGs should include multidisciplinary stakeholders, together with patients, to address the identified unmet needs in myositis care, based on growing evidence in the field.

**INTRODUCTION**

Idiopathic inflammatory myopathies (IIMs) encompass a heterogeneous group of rare autoimmune diseases showing a large degree of overlap<sup>1</sup> but also some peculiarities and differences, in particular in the setting of anti-synthetase syndrome (ASSD).<sup>2</sup> If IIMs affect adults and children, ASSD is generally an adult-onset condition,<sup>3</sup> with only few cases reported in paediatric age.<sup>4</sup> IIMs are generally characterised by the occurrence of muscle weakness and inflammation, but in ASSD, arthritis and interstitial lung disease (ILD) are more frequently

observed and frequently represent the first manifestation of the disease.<sup>5</sup> Skin can also be frequently affected by IIMs, with occurrence of lesions such as heliotrope rash, Gottron's sign/papules, skin ulcers, V-sign, Shawl sign, Hiker's feet and mechanic's hands, with these two latter considered as typical ASSD findings.<sup>6,7</sup> Additionally, IIMs are associated with cancer,<sup>8</sup> the risk of which is increased up to 5.3-fold with respect to general population in males with dermatomyositis (DM).<sup>9</sup> Thus, physicians from different specialties are involved in the care (including diagnosis, treatment and monitoring) of these patients. Even if IIMs are potentially treatable diseases, currently, mortality is hardly fourfold increased with respect to general population<sup>10</sup> and quality of life is decreased.<sup>11</sup> Considering the large number of specialists potentially involved in the daily management of these patients, as well as the different therapeutic approaches available, it is important to homogenise the different approaches, in order to establish a common shared strategy. Clinical practice guidelines (CPGs) are systematically developed statements to assist practitioner and patient decisions about appropriate healthcare for specific clinical circumstances.<sup>12,13</sup> CPGs have been proposed for IIMs, but they are sparse and not homogeneous. This work attended to identify currently available CPGs for IIMs, to provide a narrative review of their contents and to identify patients' and clinicians' unmet needs. It has been performed in the framework of the European Reference Network on rare and complex connective tissue and musculoskeletal diseases (ERN ReCONNET), a Network of centre of expertise and patients funded by the European Union's Health Program (reconnet.ern-net.eu).

## METHODS

### Objective and management of first work package of ERN ReCONNET

The aim of the first work package of ERN ReCONNET was the identification of field of application of identified CPGs, looking for potential unmet needs.

Planning and evaluation of the work was driven by regular interaction between participants of the working group during meetings (European League Against Rheumatism (EULAR) Congress 2017, American College of Rheumatology (ACR) Congress 2017, ERN ReCONNET Meeting, Pisa, 4–6 February 2018), web conferences, electronic letters and ERN Collaborative Platform (<https://webgate.ec.europa.eu>).

### Systematic literature search

Between June 2017 and February 2018, we carried out a search in PUBMED and EMBASE based on controlled terms (MeSH and Emtree), keywords of the disease and publication type (CPGs). All published articles were reviewed in order to identify existing CPGs on diagnosis, monitoring and treatment of IIMs, according to the Institute of Medicine 2011 definition (*clinical practice guidelines are statements that include recommendations intended to optimise patient care that are informed by a systematic review of*

*evidence and an assessment of the benefits and harms of alternative care options*).

The Disease Coordinators (DCs) (LC and AM) of the ERN ReCONNET for IIMs have assigned the work on CPGs to the Healthcare Providers involved. Moreover, in order to implement the list of guidelines provided by Medline and Embase search, the group performed also a hand search. A first screening of evidence-based medicine guidelines (regardless its level) was performed on the basis of their title and abstract (systematic search and hand search). A discussion group was set for the effective inclusion and evaluation of the existing CPGs, in order to identify the unmet needs.

Here is the search strategy: *Medline (Pubmed)*: ("polymyositis" [MeSH Terms] OR "polymyositis" [All Fields] OR "dermatomyositis" [MeSH Terms] OR "dermatomyositis" [All Fields] OR "myositis" [MeSH Terms] OR "myositis" [All Fields] OR ("idiopathic" [All Fields] AND "inflammatory" [All Fields] AND "myopathies" [All Fields]) OR "idiopathic inflammatory myopathies" [All Fields]) AND ("Practice Guideline" [Publication Type] OR "Practice Guidelines As Topic" [MeSH Terms] OR Practice Guideline [Publication Type] OR "Practice Guideline" [Text Word] OR "Practice Guidelines" [Text Word] OR "Guideline" [Publication Type] OR "Guidelines As Topic" [MeSH Terms] OR Guideline [Publication Type] OR "Guideline" [Text Word] OR "Guidelines" [Text Word] OR "Consensus Development Conference" [Publication Type] OR "Consensus Development Conferences As Topic" [MeSH Terms] OR "Consensus" [MeSH Terms] OR "Consensus" [Text Word] OR "Recommendation" [Text Word] OR "Recommendations" [Text Word] OR "Best Practice" [Text Word] OR "Best Practices" [Text Word]). *Embase*: ('myositis'/exp OR 'idiopathic inflammatory myopathy' OR 'inflammatory myopathy' OR 'muscle inflammation' OR 'myopathy, inflammatory' OR 'polymyositis'/exp OR 'fibromyositis' OR 'polymyositis' OR 'dermatomyositis'/exp OR 'dermatomyositides' OR 'dermatomyositis' OR 'polymyositis arthropathica') AND ('practice guideline'/exp OR 'practice guideline' OR 'practice guidelines'/exp OR 'practice guidelines' OR 'clinical practice guideline'/exp OR 'clinical practice guideline' OR 'clinical practice guidelines'/exp OR 'clinical practice guidelines' OR 'clinical practice guidelines as topic'/exp OR 'clinical practice guidelines as topic' OR 'guideline'/exp OR 'guideline' OR 'guidelines'/exp OR 'guidelines' OR 'guidelines as topic'/exp OR 'guidelines as topic' OR 'consensus development'/exp OR 'consensus development' OR 'consensus development conference'/exp OR 'consensus development conference' OR 'consensus development conferences'/exp OR 'consensus development conferences' OR 'consensus development conferences as topic'/exp OR 'consensus development conferences as topic' OR 'consensus'/exp OR 'consensus' OR 'recommendation' OR 'recommendations') AND [embase]/lim NOT [medline]/lim.

### Inclusion criteria and CPGs identification

CPGs were included if<sup>12 13</sup>:

- Their development was aimed at assisting practitioners' and patients' decisions about appropriate healthcare for patients with IIMs and ASSD.
- They were based on a systematic review of evidence.
- They were written in English.

All papers identified during the literature search were screened for eligibility by two evaluators and by DCs based on title and abstract assessment. Only papers considered eligible by at least one evaluator were included. Papers selected after the first round were then further assessed in order to verify their actual status of CPGs. In case of disagreement in the decision, a further round of discussion involving participants has been performed, in order to uniform the evaluation reaching the 100% of consensus.

Physicians' and patients' unmet needs were delineated by focus groups conducted during the ERN meeting followed by written exchange during the redaction process of the present manuscript. Patients were a 51-year-old woman with bachelor's degree, a 45-year-old man with bachelor's degree and a 39-year-old woman with higher level of education.

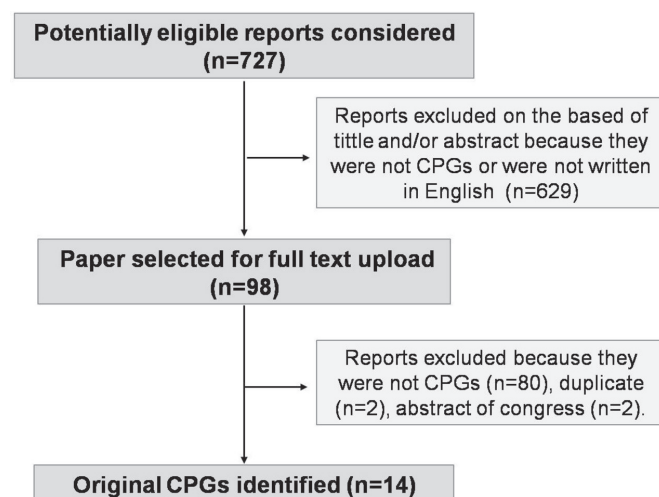
## RESULTS: STATE OF THE ART ON CPGs

### Identification of existing CPGs

The systematic literature search provided a total of 727 citations. Title and abstract evaluation identified 98 papers suitable for full-text upload and review. After full-text revision, we identified 14 original papers published between 1996 and 2017<sup>14-32</sup> (figure 1).

In the following sections, these papers will be thus defined as CPGs for practical purpose.

This is the first review providing an overview of currently available CPGs for IIMs whose characteristics and contents are resumed below and in tables 1 and 2.



**Figure 1** Flow chart constructed from Pubmed, Embase and national databases. CPGs, clinical practice guidelines.

### CPGs CHARACTERISTICS

CPGs were written (first and/or last authors) by rheumatologists (n=3), neurologists (n=7), dermatologists (n=3) and pulmonologists (n=1). In four CPGs, co-workers from different specialties than main authors were also involved. No CPGs involved clinicians from more than two different specialties. No CPGs involved patients.

Five CPGs were dedicated to IIMs, while nine covered a broader spectrum of diseases (including IIMs).

Nine CPGs targeted IIMs as a whole (juvenile and adult) although in four of the latter,<sup>14-16 23</sup> authors did not state whether the recommendation applied to adult and/or juvenile IM. One paper targeted specifically juvenile IIMs and four specifically adults IIMs.

The majority of CPGs (n=13) focused on therapeutic questions including overall treatment, use of intravenous globulin, management of lung and cutaneous disease. Others addressed disease measurements (tools for clinical assessment and improvement definition), associated conditions (cancer, pregnancy) and overall management. In seven CPGs, evidence was systematically searched and discussed (evidence-based CPGs). In the others, the method for evidence search was not provided ('eminence-based' CPGs). There were no specific recommendations focusing on ASSD.

### CPGs CONTENT

The covered points are as follows. (1) Global management of juvenile dermatomyositis (DM), the most frequent IIMs in children,<sup>20</sup> and of IIMs in general.<sup>19 23 31</sup> (2) The role of physical therapy in IIMs, focusing on the evaluation of disease-specific quality indicators, outcome measures and guidelines. The establishment of a physical rehabilitation path, in association to the pharmacological treatments, is crucial to obtain the best improvement possible in these patients.<sup>15</sup> (3) Screening of cancer, one of the leading causes of death in IIMs.<sup>32</sup> (4) Use of intravenous immunoglobulin (IVIG), an expensive treatment that should be reserved only to specific clinical conditions.<sup>14 16 17 21 29</sup> (5) Cutaneous manifestations which are debilitating complications of various IIMs,<sup>22</sup> in particular in juvenile forms.<sup>20</sup> (6) ILD-related treatment,<sup>28</sup> the main impacting prognostic factor in IIMs in general and in ASSD in particular. (7) Pregnancy planning and management, in a field in which juvenile forms are common and young people in childbearing age can be affected by these diseases.<sup>18</sup>

Recommendations of available CPGs are presented in table 2 and notably stated that: (1) extramuscular involvements should be assessed; (2) corticosteroids and methotrexate or azathioprine are first-line therapies of IIMs; (3) IVIG is a treatment of resistant-DM that may be also used in other resistant-IIMs; (4) physical therapy and sun protection (in patients with DM) are part of the treatment; (5) tumour screening for patients with DM include imaging of chest, abdomen, pelvis and breast (in woman) along with colonoscopy (in patients over 50 years); (6)



**Table 1** CPGs general characteristics

Author	Date	Target	Main authors specialty	Other specialty involved	Based on	Scope
Drake <i>et al</i> <sup>19</sup>	1996	IIMs as a whole	Dermatology	None	Evidence (grade C–D)	Treatment (overall)
Bril <i>et al</i> <sup>16</sup>	1999	IIMs as a whole	Neurology	None	Eminence	Treatment (IVIG)
Doria <i>et al</i> <sup>18</sup>	2004	Adult IIMs	Rheumatology	Neurology	Eminence	Pregnancy
Feasby <i>et al</i> <sup>21</sup>	2007	IIMs as a whole	Neurology	None	Evidence (grade B–D)	Treatment (IVIG)
Alexanderson <i>et al</i> <sup>15</sup>	2007	IIMs as a whole	Rheumatology	Physical therapy	Eminence	Disease measurement (tools)
Donofrio <i>et al</i> <sup>17</sup>	2009	Adult IIMs	Neurology	None	Evidence (grade B–D)	Treatment (IVIG)
Hengstman <i>et al</i> <sup>23</sup>	2009	IIMs as a whole	Neurology	None	Eminence	Treatment (overall)
Titulaer <i>et al</i> <sup>32</sup>	2011	IIMs as a whole	Neurology	None	Evidence (grade C–D)	Tumour screening
Patwa <i>et al</i> <sup>29</sup>	2012	Adult IIMs	Neurology	None	Evidence (grade B–D)	Treatment (IVIG)
Sunderkotter <i>et al</i> <sup>31</sup>	2016	IIMs as a whole	Neurology	Dermatologist	Eminence	General management
Enk <i>et al</i> <sup>14</sup>	2016	IIMs as a whole	Dermatology	None	Eminence	Treatment (IVIG)
Fujimoto <i>et al</i> <sup>22</sup>	2016	IIMs as a whole	Dermatology	None	Evidence (grade C–D)	Treatment (skin disease)
Morrisset <i>et al</i> <sup>28</sup>	2016	Adult IIMs	Pneumology	None	Eminence	Lung disease
Enders <i>et al</i> <sup>20</sup>	2017	Juvenile IIMs	Rheumatology	None	Evidence (grade B–D)	General management

Evidence was graded according to Grading of Recommendations Assessment, Development and Evaluation Working Group 2007 (<http://www.gradeworkinggroup.org/>)

CPG: clinical practice guideline; IIM: idiopathic inflammatory myopathy; IVIG: intravenous immunoglobulin;

disease activity and damages should be monitored using standardised and validated tools.

## UNMET NEEDS

CPGs that are currently available do not cover the entire spectrum of IIMs needs. From the clinical point of view, we think that the identification of uncovered areas could be useful for the establishment of additional CPGs. From the scientific point of view, by considering that CPGs are recommendations based on scientific evidences, some of the unmet needs could indicate areas that deserve future research. Finally, it will be important to critically review the content of existing CPGs, in order to identify their strength and weakness, looking to their improvement, as planned in the next ReCONNET work packages.

### Clinicians' unmet needs

Even if several issues have been addressed, other relevant points are currently not or ineffectively targeted by CPGs IIMs that can be summarised as follows (table 3):

#### Unmet needs related to the stakeholders of CPGs development

- The lack of large multidisciplinary working group is the first transversal and crucial problem of available CPGs. Indeed, considering that patients with IIMs have multiorgan involvement with different manifestations prevalence and severity levels, CPG for their

optimal management cannot be developed without a larger number of different organ specialists as stakeholders. In this regard, a collaboration of ReCONNET with other ERNs concern by IIMs (EURO-NMD and ERN LUNG) has been planned.

- Second, as IIMs are chronic diseases, patients' perspective is mandatory for the improvement of daily quality of care. This issue will be overcome by initiatives notably started by the OMERACT (independent initiative of international health professionals interested in outcome measures in rheumatology).<sup>33</sup>

#### Unmet needs related to scope and target of the CPGs

- No CPGs are currently available for diagnosis. Early intervention may be associated with a better outcome<sup>34</sup> but treatment is often delayed or not appropriated, because IIMs can be misdiagnosed as numerous other conditions including non-inflammatory myopathies<sup>34</sup> and other inflammatory diseases (such as rheumatoid arthritis<sup>35</sup> or ILDs with autoimmune features<sup>36</sup>). On the other hand, mimickers of treatable IIMs include numerous non-inflammatory myopathies and inclusion body myositis.<sup>37</sup> Although being an important milestone in the field, the recently published EULAR/ACR IIMs classification criteria did not match the definition of CPGs and were thus not included in this review. Yet, even considering

**Table 2** Scope and key recommendations of available CPGs for IIMs

Scope	First author, date of publication (ref)	Key recommendations of the CPG
General management	Sunderkotter, 2016 <sup>31</sup>	<ul style="list-style-type: none"> <li>▶ All patients with IIMs should undergo a pulmonary function test and—in case of pathological findings—further pulmonology workup.</li> <li>▶ Tumour screening in DM with anti-TIF1-<math>\gamma</math> include 18-fluoro-deoxyglucose-positron emission tomography/CT or CT of the thorax and abdomen in combination with a gynaecological/urological examination is recommended (no strategy is provided in other case by the CPG).</li> <li>▶ Treatment of IIMs includes:               <ul style="list-style-type: none"> <li>– CS (intravenous if severe, oral if not).</li> <li>– AZA (for adult) and MTX (for children) when: (1) severe IIMs, (2) impossibility to reduce CS dosage below the ‘Cushing threshold’ after 3 months.</li> <li>– IVIG is recommended in patients unresponsive to CS+AZA.</li> <li>– Non-pharmacological measures: regular physical therapy, sun protection (for patients with DM).</li> </ul> </li> </ul>
	Enders, 2017 <sup>20</sup>	<ul style="list-style-type: none"> <li>▶ All children with suspected IIMs should be referred to a specialised centre. High-risk patients (as defined in the CPG) need immediate/urgent referral.</li> <li>▶ Assessment of organ involvement (muscle, skin, lung, heart), calcinosis and antibodies (see CPG for details) is recommended for all children with IIMs.</li> <li>▶ Disease activity, damages and health status should be monitored in a standardised way.</li> <li>▶ Juvenile IIMs treatment includes:               <ul style="list-style-type: none"> <li>– Sun protection, exercise programme.</li> <li>– First line: high-dose CS and MTX.</li> <li>– If failure (considered within the first 12 weeks): topical TACRO/ CS (if localised skin disease), CsA or MMF (if intolerance to MTX), IVIG as adjunct or RTX as adjunct or CYC or antitumour necrosis factor therapies (if resistance).</li> </ul> </li> </ul>
Treatment (overall)	Drake, 1996 <sup>19</sup>	<ul style="list-style-type: none"> <li>▶ Non-pharmacological treatments of IIMs include physical therapy, photoprotection and adequate nutrition.</li> <li>▶ Pharmacological treatments of IIMs include CS (topical and systemic); antimalarial and CS-sparing agents (no limitative list provided in the CPG).</li> <li>▶ Calcinosis treatments include medical (diphosphonates, aluminium hydroxide, probenecid, colchicine and low-dose warfarin) and surgical management.</li> </ul>
	Hengstman, 2009 <sup>23</sup>	<ul style="list-style-type: none"> <li>▶ First line of IIMs is prednisone (1 mg/kg) and AZA or MTX.</li> <li>▶ Second line, in case of severe pulmonary involvement, is CsA or TACRO or CYC.</li> <li>▶ Second line, in case of no severe pulmonary involvement, is IVIG.</li> <li>▶ Third line is RTX or TACRO or MMF or CsA or CYC.</li> </ul>
Treatment (IVIG)	Bril, 1999 <sup>16</sup>	▶ IVIG is favourably recommended for the treatment of DM and recommended as a last resort for the other IIMs.
	Feasby, 2007 <sup>21</sup>	<ul style="list-style-type: none"> <li>▶ IVIG is recommended as an adjunctive treatment for DM who did not adequately respond to other immunosuppressant medications (such as CS, MTX or AZA).</li> <li>▶ IVIG may be considered as an adjunctive treatment option for PM who failed to respond to first-line therapies.</li> <li>▶ IVIG is not recommended for IBM.</li> </ul>
	Donofrio, 2009 <sup>17</sup>	<ul style="list-style-type: none"> <li>▶ IVIG therapy is recommended as add-on treatment in refractory IIMs.</li> <li>▶ IVIG is not recommended for IBM.</li> </ul>
	Patwa, 2012 <sup>29</sup>	<ul style="list-style-type: none"> <li>▶ IVIG may be considered for the treatment of non-responsive adult DM.</li> <li>▶ Evidence is insufficient to support or refute the use of IVIG in treating IBM and PM.</li> </ul>
	Enk, 2016 <sup>14</sup>	Severe forms of DM, PM and IBM are considered by the authors as indication of IVIG, as first-line treatment (in fulminant course, severe myolysis or paralysis) or second-line treatment (in other cases), with continuation of immunosuppressive therapy.
Pregnancy	Doria, 2004 <sup>18</sup>	<ul style="list-style-type: none"> <li>▶ Patients with IIMs should be correctly informed on the risk of becoming pregnant.</li> <li>▶ Pregnancies should be planned when IIMs is in remission</li> <li>▶ Patients with IIMs should be regularly monitored during gestation and postpartum by a multidisciplinary team.</li> <li>▶ In the case of disease relapse, treatment has to be started as soon as possible.</li> <li>▶ IIMs treatment in pregnant patients is: CS (1 mg/kg/day until thenormalisation of serum creatine kinase levels). If insufficient response: CsA, AZA, plasma exchange or IVIG.</li> </ul>

Continued

Table 2 Continued

Scope	First author, date of publication (ref)	Key recommendations of the CPG
Disease measurement (tools)	Alexanderson, 2007 <sup>15</sup>	<ul style="list-style-type: none"> <li>▶ Disease activity and disability should be measured at disease onset, at 3, 6 and 12 months, and then at least once a year.</li> <li>▶ Only valid and reliable clinical outcome measures should be used (such clinical outcome measures were not available at the time of the publication of the CPG).</li> </ul>
Tumour screening	Titulaer, 2011 <sup>32</sup>	<ul style="list-style-type: none"> <li>▶ Patients with DM should have CT-thorax/abdomen, ultrasound of the pelvic region and mammography in women, ultrasound of testes in men under 50 years and colonoscopy in men and women over 50.</li> <li>▶ If primary screening is negative, repeat screening after 3–6 months and screen every 6 months up till 4 years.</li> </ul>
Extramuscular involvement (skin)	Fujimoto, 2016 <sup>22</sup>	<ul style="list-style-type: none"> <li>▶ For calcinosis in IIMs: low-dose warfarin, aluminium hydroxide gel, diltiazem hydrochloride, probenecid or bisphosphonate are recommended as an option. Surgical treatment is also recommended as an option.</li> <li>▶ For panniculitis in IIMs: CS is recommended. If no response, immunosuppressants such as CsA, MTX and AZA are recommended as an option.</li> </ul>
Extramuscular involvement (lung)	Morrisset, 2016 <sup>28*</sup>	<ul style="list-style-type: none"> <li>▶ In acute or severe IIMs–ILD: high-dose steroids+CYC or RTX or CsA or TACRO is recommended.</li> <li>▶ In chronic or mild to moderate IIMs–ILD: steroids+MMF or AZA is recommended.</li> <li>▶ In case of failure: it is recommended to switch agent or consider combination of agents, or consider IVIG or consider transplantation referral.</li> </ul>

AZA, azathioprine; CPGs, clinical practice guidelines; CS, corticosteroid; CYC, cyclophosphamide; CsA, ciclosporin A; DM, dermatomyositis; IBM, inclusion body myositis; IIM: idiopathic inflammatory myopathy; ILD: interstitial lung disease; IVIG, intravenous immunoglobulin; MMF, mycophenolate mofetil; MTX, methotrexate; PM, polymyositis; RTX, rituximab; TACRO, tacrolimus.

these classification criteria,<sup>38</sup> lung, joint and skin (other than DM rash) involvements are not taken into account,<sup>39</sup> which is an issue especially for diagnosis of ASSD, as previously suggested by the American, European Network of Antisynthetase Syndrome collaborative group.<sup>2 36 39–41</sup> This unmet need will be fixed in next year thanks to CLASS project addressed to the establishment of next EULAR/ACR classification criteria of ASSD.

▶ Management of extramuscular organ involvement other than skin is not appropriately targeted by current available CPGs. Of note, four papers address this important point, but have not been included in this review as they are based on expert consensus and thus do not match criteria for CPGs.<sup>24–27</sup> Lung disease, one of the most frequent manifestations of IIMs and major cause of death<sup>10</sup> is only addressed by one CPG.<sup>28</sup> No CPG currently address heart, joints

Table 3 Synthesis of the physicians' unmet needs

Unmet need related with	Unmet needs	Description
Stakeholders	Patients' preference	None of the CPGs are based on patients preference.
	Multidisciplinary working group	None of the CPGs involved >2 different specialities.
CPGs scope and targets	Diagnosis	None of the CPGs provide a integrated strategy for diagnosis.
	Extramuscular involvement (other than skin)	Only one eminence-based CPG cover diagnosis and management of ILD. No CPG specifically address management of heart, joints and gastroenterological manifestations.
	Management by subgroups of IIMs	None of the CPG is personalised by subgroup of inflammatory myopathies below than DM versus other IIMs.
	Activity and damage assessment	Only one eminence-based CPG specifically addresses disease activity and damages assessment.
	Life or organ-function threatening complications	None of the CPG cover management of life or organ-function threatening complications.
	Comorbidities	None of the CPG cover comorbidities prevention, diagnosis and treatment.
	Transition management	None of the CPG targets transition management.

CPGs, clinical practice guidelines; DM, dermatomyositis; IIM: idiopathic inflammatory myopathy; ILD: interstitial lung disease.

and gastroenterological manifestations, which are yet source of handicap and/or increased mortality.<sup>42–44</sup>

- ▶ IIMs are a heterogeneous group of diseases. The clinical manifestations, autoantibody profile and muscle histology can be used to distinguish patient subgroups (extending their spectrum far beyond the subdivision between DM and other IIMs) with fairly homogeneous patterns of complications, treatment responses and outcomes. Yet, none of the CPGs identified in this review were specifically dedicated to one of these subgroups.
- ▶ Accurate monitoring of muscle inflammatory activity and damages are mandatory in order to meet the subtle threshold point between undertreatment and overtreatment. This challenge has been taken up by only one CPG. The recently published EULAR/ACR criteria for clinical response in adult and juvenile IIMs<sup>45–46</sup> is an important step in this field. Yet, they were not included in this review because they did not match the definition of CPGs.
- ▶ Patients with IIMs may face life or organ-function threatening complications such as severe rapidly progressive ILD, myocarditis, pneumomediastinum, pulmonary hypertension, respiratory failure, prone position complication due to severe muscle weakness dysphagia and ischaemic ulcerative colitis. These situations require peculiarly prompt diagnosis and management. Yet, no CPG currently cover these points.
- ▶ Side-effects of IIMs treatments and comorbidities (such as infections, osteoporosis, cardiovascular events and high-risk pregnancy) are major causes of morbidity and mortality in patients with IIM.<sup>10–47</sup> Prevention and management of these associated conditions are currently not or only insufficiently targeted by CPGs.
- ▶ Transition management. IIMs encompass all ages with different disease phenotypes and different patients' needs, in particular in the transition period. Patients' approach of adult and paediatric clinicians is generally different, and this may be an additional serious problem to be considered.

### Patients' unmet needs

This paragraph intends to highlight the unmet needs of the IIMs European community. The content of this paragraph has been carefully but hardly realised collecting the voices and the points of view of the patients affected by the disease. The rarity of the disease and the scarcity of patient organisations made this work harder than expected. IIMs spectrum disorders encompass a large number of conditions with a very heterogeneous range of manifestations, not limited to muscle involvement. On this basis, it is evident that patients' needs are very different and deeply influenced by disease presentation pattern. Furthermore, also the diagnosis is not always easy to reach and this adds further uncertainty to patients, with a large number of doubts about

the carried disease: 'I do not understand what I have?', 'What is happening to me?', 'What should I expect from the future?'. Even if crucial, these questions have not always an answer in IIMs or, even worse, they have different answers according to the different referring specialist. Patients experience a lack of harmonisation in the medical approach and a scarce involvement in developing a common path. Being the diseases so heterogeneous, each patient would bring its own experiences and suggestions adding further useful information and improving the care of these conditions. Unfortunately, only few clinicians are aware that patients need more direct support and attention: a continuous process of discussion and dialogue with physicians could be surely relevant to this purpose. Furthermore, patients refer that they need more personalised assistance from the national health systems, in term of social demands and attention. The lack of harmonisation of assistance creates differences in the European Countries and ERN ReCONNET is perceived to possibly fill this gap.

### CONCLUSIONS

We identified the currently IIMs available CPGs, showing a large area of unmet needs. A further effort is necessary and it has been planned by the ERN ReCONNET in order to evaluate the intrinsic validity and applicability of available recommendations. The standardisation and sharing of CPGs, from diagnosis to treatment, are crucial to really improve the prognosis of these patients. We need therefore to work intensively together with patients focusing on the global improvement of their daily life that represents a shared objective of patients and clinicians. We propose that future CPGs should include multidisciplinary stakeholders, together with patients, to address the identified unmet needs in myositis care, based on growing evidences.

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## Correction: Idiopathic inflammatory myopathies: narrative review of unmet needs in clinical practice guidelines

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This article has been corrected since it first published. The authors would like to notify that title of the article has been changed to:

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